A Cephalometric Evaluation of the Cranial Base in Microcephaly*

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Introduction

Anatomical research in recent years indicates that the skull base is a critical region in the coordination of growth of the neural and facial skeletons. Only a few studies have dealt with aberrant growth of this region. Further investigations could lead to a better understanding of the influence of the cranial base on the dentofacial complex. Todd[1] and others[2-4] have shown that one area of disharmonic growth will cause subsequent abnormalities in regions and structures correlated with it. Many dentofacial disharmonies exist, however, without disturbances of the skull base.

In an effort to clarify normal growth processes of the cranial base, microcephalics, who have an obvious disharmony of growth in a region contiguous to the skull base, have been studied. It would seem that such a gross disproportion in growth of the neural mass should have some effect on the cranial base. All reports in the literature, however, are to the contrary. The cranial base is said to be unaffected by a reduction of cranial capacity.

The current study was undertaken in order to apply a method of measuring cranial capacity *in vivo* to the diagnosis of microcephaly. It was also hoped to determine the variability, if any, of the microcephalic cranial base with the normal cranial base, as well as to determine possible clinical applications.

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Materials and Methods

Thirty-five subjects, medically diagnosed as microcephalic, were selected from the Fernald School, Waltham, Massachusetts, under the supervision of the Department of Neurology, Massachusetts General Hospital, Boston. The subjects were varying ages and of both sexes. The following records were taken: 1) lateral cephalograms, 2) posteroanterior cephalograms, 3) wrist roentgenograms on all subjects under fifteen years of age, 4) profile and front view photographs, 5) dental casts (unless totally edentulous), and 6) finger and hand (palm) prints of interrelated subjects. All of the above records were not used in this particular study but were taken for concurrent research and possible future studies. Twelve of the original subjects were eliminated because of incomplete or inaccurate records as a result of severe mental retardation and/or poor cooperation. One Mongoloid and two dwarfs were also eliminated. One subject had superimposed pathology in the region of the nose and eyes and was, therefore, eliminated from the study. The remaining nineteen subjects were used for this study.

The cephalometer was adjusted to conform to the method described by Björk.[5] The focus-median plane distance was 180 cm and the median plane-film distance was as close to 9 cm as possible.

Tracings were made from both lateral and P-A cephalograms on matte acetate tracing film. A typical tracing of the lateral cephalogram is illustrated in Figure 1. The following reference points were used: nasion (N), sella (S), basion

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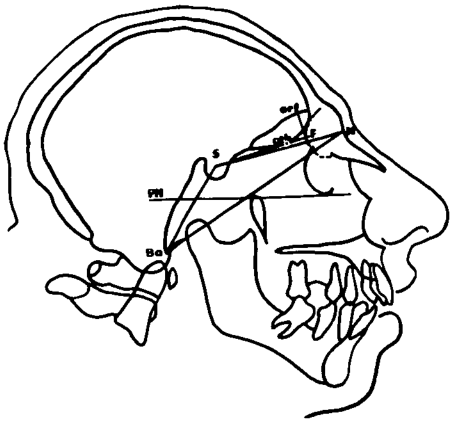

Fig. 1 Typical tracing of a cephalogram of a microcephalic. Orientation to the Frankfort Horizontal Plane (FH) is indicated.

(Ba), ethmoidal (eth) (the lowest median point of the contour of the anterior cranial fossa corresponding to the cribriform plate of the ethmoid), orbital roof (orf) (the uppermost point in the roof of the orbital cavity) and frontal bone (F) (a point along the N-S line indicating the internal contour of the frontal bone; this is marked by projecting the distance from nasion of the nearest point on the internal contour of the frontal bone).

The following reference lines were measured to the nearest 0.5 mm: N-S, N-F, S-F, S-Ba, eth-NSL (the perpendicular distance from ethmoidal to N-S line; the value of this line is positive when ethmoidal is situated below the N-S line and negative when ethmoidal is situated above N-S line) and orf-NSL (the perpendicular distance from the uppermost point of the roof of the orbital cavity to N-S line).

The cranial volume was measured from the lateral and P-A cephalograms in accordance with the technique developed by Neihoff and Haach.[6]

All measurements, linear as well as angular, were read to the nearest half of a millimeter or to the nearest half of a degree, respectively. The data obtained were then subjected to suitable statistical analysis. This will be discussed subsequently.

Findings

The normal means and ranges for American children and young adults have been reported.[7] All subjects used in this study were two standard deviations or more below the normal mean. In addition, each subject had been medically diagnosed as microcephalic. No differentiation was made as to sex or etiological factors.

The control group for the cranial base measurements in this investigation was taken from a study of Björk[8] and one by Stramrud.[9] These investigators used identical cephalometric techniques. Stramrud's sample consisted of 464 Danish males ranging from three to twenty-five years of age. Except for the youngest group, thirty or more individuals were in each group. Björk's sample consisted of 243 twelve-year-old Swedish males who were re-evaluated at age twenty. The mean and standard deviations for the cranial base reported by these investigators are well within one standard deviation of each other and generally are accepted as representative of the normal population. The cephalometric technique employed in our study was identical to the studies cited[8,9] so that correction for distance was unnecessary.

Age is an important consideration in studies involving the cranial base. Subjects over the age of twenty would be ideal, since growth of the structures comprising the cranial base is virtually completed by the age of twenty. The number of available microcephalics, however, was limited. It was impossible to gather a sufficient sample without including younger subjects. A minimum age requirement of twelve years was established to assure that growth was

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TABLE I
Comparison of Sample to Normal

	Normal		Sample		±
	Mean	S.D.	Mean	S.D.	
Age			32.4	16.8	
Head Circ.	55.6	1.8	49.1	2.3	15.7*
Cranial Vol.			1016.8	198.4	
N-S	73.7	3.7	66.5	5.1	8.5*
N-F	16.4	2.7	14.3	2.8	3.5*
S-F	57.3	3.4	52.3	5.0	6.4*
S-Ba	48.5	2.7	42.6	3.5	9.5*
eth-NSL	2.4	2.2	.3	2.1	5.3*
orf-NSL	14.4	1.5	14.6	1.5	.6
N-Ba	112.0	4.4	100.6	6.3	11.3*
N-S-Ba	131.2	4.8	133.8	4.5	2.4**
eth-S-Ba	128.1	4.5	134.7	6.0	6.4*

* Significance greater than .01
** Significance greater than .05

completed between the foramen caecum and sella turcica. The sample consisted of six subjects below the age of twenty years and thirteen over the age of twenty. Table I compares the means and standard deviations of the total sample with the normal. A t-value was computed and recorded.

The thickness of the frontal bone (N-F) was closely correlated with the distance of the cribriform plate (eth) to the nasion-sella line (NSL). As the frontal bone thickened, the distance between ethmoidale and the nasion-sella line decreased. Another interesting correlation is the ethmoidale to NSL and the eth-S-Ba angle. As the distance from the cribriform plate to the nasion-sella line decreased, the internal cranial base angulation (eth-S-Ba) increased.

Table II lists all correlations tabulated for each measurement using all nineteen.

The subjects were further divided into two groups: 1) cranial capacity greater or less than 1100 cc and, 2) cranial circumference greater or less than 50 cm. Nonsignificant t-scores were obtained and, therefore, for purposes of brevity are not reported here.

TABLE II
Significant Correlations

1	9	.5247**
2	3	.7466*
2	4	.5541**
2	10	.4496**
4	6	.8466*
4	10	.8846*
5	8	.6789*
6	10	.6889*
7	10	.6237*
8	12	.5528**
9	10	.4675**
11	12	.7462*

1=Age 5=N-F
2=Head Circum. 6=S-F
3=Cranial Vol. 7=S-Ba
4=N-S 8=eth-NSL
9=orf-NSL
10=N-Ba
11=N-S-Ba
12=eth-S-Ba

* Significance greater than .01
** Significance greater than .05

Discussion

In reviewing the literature it becomes apparent that the mechanism of growth of the cranial base is not fully understood. Furthermore, the influence exerted by the growing cranial base on

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surrounding structures as well as the influence of the surrounding structures (brain and face) on the growing cranial base is not known. Brodie, Jr.,[10] and Pruzansky and Lis[11] realized that the study of abnormal growth processes frequently clarifies some of the mechanisms of the normal growth process. This investigation was undertaken in an effort to understand more clearly the normal by a study of the abnormal.

In order to establish that the sample is abnormal, the accurate diagnosis of microcephaly is of paramount importance. Because of the variation in the skull thickness and integumental tissues of individuals, circumferential measurements of the cranium have been considered to be only approximations of the cranial capacity. Microcephaly has recently been more accurately termed "micrencephaly" by some investigators[12-14] It would appear logical that the measurement of cranial capacity in living subjects would result in a more accurate determination of "micrencephaly." In other words, microcephaly as a pathological entity would be more accurately defined and diagnosed by cranial volume measurement than by measurement of cranial circumference.

The cranial base of the microcephalic has never been subjected to close scrutiny. Measurements directly on a small number of microcephalic skulls (less than ten) yielded reports of a "normal" cranial base and "normal" facial development.[15-17] An *in vivo* cephalometric study of the cranial base of microcephalics has never been reported. The results of the present investigation indicate a significant decrease in size of the cranial base in microcephaly. Each segment of the cranial base (N-S or S-Ba or F-S) was also found to be significantly shorter. Individual subjects frequently had one portion or another of the cranial base within the normal range but in almost all instances it was found to be at the lower end of the normal range. In examining microcephalic skulls, Scott[16] reported that the length of the cranial base usually was within the normal range; he did not note that they were mostly at the lower end of the range. In the current study the S-F distance in eight of nineteen subjects was within the normal range. The mean average of the total sample, however, resulted in a t of 6.31. The same was found to be true of the other segments of the cranial base (N-S, $t = 6.5$; S-Ba, $t = 9.2$).

An explanation for the shorter cranial base is not readily apparent. Microcephaly is reported by some investigators as resulting from a monorecessive gene. It is possible that the genetic determination for growth of the cranial base may be affected, as is the genetic potential for brain size. Then again, microcephalics are known to be slightly smaller in stature; hence, the shorter cranial base may simply reflect the decreased body size. Whether or not the slightly-dwarfed body is genetically influenced is not known.

The theory of the functional matrix would also seem to offer a logical explanation. Moss[18] stated, "It is the growth of the functional matrix (i.e., those tissues supported or protected by functionally-related skeletal elements) which furnishes the primary growth force; the bones respond secondarily." Ford[19] wrote, "The part of the cranial base from foramen caecum to sella is so intimately related to the frontal lobes of the brain that it would be surprising if it did not have the same growth rate as the brain itself." These two statements would suggest that the internal cranial base of the microcephalic would be responsive to the retarded growth activity of the brain and should, in some manner, reflect the smaller neural mass. Moss,[20] however, gave another explanation for what he believed was a *normal*

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cranial base in the microcephalic. The structures at the base of the brain are related to ophthalmic and olfactory function, which are normal in the microcephalic. Therefore, the base of the brain is normal, he stated, and so too is the cranial base upon which it lies.

Baume,[21] in noting that the cranial base was preformed in cartilage, stated that cartilage was highly resistant to mechanical pressures and that the cranial base grew to a genetically predetermined size and shape. If the theory of the functional matrix were accepted, it is difficult to reconcile this with the fact that the brain mass could influence the ultimate size of the underlying cartilaginous base.

The posterior cranial base (S-Ba) has been described as following the skeletal growth rate. Since the activity of the spheno-occipital synchondrosis continues into adolescence, it is believed that this area of the cranial base is more intimately related to the growth of the splanchnocranium (facial skull) than to that of the brain. It is well established that the growth of the neural mass is virtually completed by eight years of age. The splanchnocranium of the microcephalic is said to be normal; therefore, it is surprising to find the posterior cranial base (S-Ba) considerably shortened ($t = 9.50$). The size of the brain must exert a greater influence on this portion of the cranial base than heretofore believed.

The effect of the shorter cranial base on the face should be further investigated. The face of the microcephalic, visually, appears larger than normal. This, undoubtedly, is due to the contrast it presents with the small cranium. Korkhaus[3] reported the cranial base and face of the microcephalic as normal. He did not support statistically this statement. With a shorter cranial base we could, therefore, expect to find changes in the microcephalic face. The possibility of compensatory growth of the facial structures, however, must be considered. Björk and Björk[22] reported a case in normal face, even though alterations existed in the cranial base. Further investigation in this area could provide important information on facial growth.

The external cranial base angulation (N-S-Ba) of the microcephalic was found to have a range of 125.0° to 140.0° and a mean value of 133.8±4.5. The t value was 2.4. This is considered to be moderately significant at the one per cent level and would seem to indicate a slight platybasia of the cranial base. This is supported by the mean internal cranial base angulation (eth-S-Ba) of 134.7° (normal: 128.1°) which gave a t value of 6.4 (Table I).

Björk[8] and Stramrud[9] established norms for the measurement of the eth to NSL. Stramrud reported norms for orf to NSL. In the microcephalic, the orf-NSL distance is within the normal range, but the eth-NSL distance is altered significantly ($t = 5.3$).

If we assume that the N-S line has remained stable in microcephalics as is reported for normals,[8,9,23] the discrepancy of eth-NSL distance would appear to be the result of decreased inferior displacement of the cribriform plate. An interesting significant correlation, however, was found between the frontal bone thickness (N-F) and the eth-NSL distance (Table II). As the thickness of the frontal bone increased, the distance of the eth-NSL shortened. This indicates that nasion in the microcephalic migrates inferiorly and that the nasion-sella line is not stable.

Another correlation, eth-NSL and eth-S-Ba, was also found to be significant. As the distance of eth-NSL decreased, the angulation eth-S-Ba increased, or platybasia of the cranial base occurred. This could indicate that the apparent lack of downward positioning

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of the cribriform plate may not be entirely due to an inferior movement of the nasofrontal suture. Since the roof of the orbit (orf) is related to the eye, which is considered normal in the microcephalic, an inferior movement of nasion should be reflected in the distance of orf-NSL line. Since this is not the case, the correlation of eth-NSL to N-F is questionable. It must be noted that the limited number of subjects used in this study does not permit the strictest interpretation of these data.

Correlations were computed for each series of measurements in which certain obvious correlations appear (Table II). For example, cranial volume to head circumference (2-3) and segments of the cranial base length to the total cranial base length (4-6, 6-10, 4-10) all displayed significant correlations. The length of the cranial base was more closely correlated to cranial circumference than it was to cranial volume. The angulation of N-S-Ba was closely correlated to eth-S-Ba angulation. The N-Ba distance was closely correlated to the N-S-Ba angulation. The cranial volume, however, was not found to be correlated to any changes in the cranial base. This is somewhat surprising, since one might expect changes in the cranial base in direct relation to the size of the neural mass.

The thickness of the frontal bone (N-F) is within the normal range for subjects under twenty years of age ($t = 1.1$). Among those over twenty years of age, the frontal bone thickness appears slightly reduced in size ($t = 2.1$). Due to the small number of subjects as well as to the lack of differentiation in sexes, the significance of this finding is questionable.

The microcephalic cranial base is significantly altered from the normal. An intensive study of the microcephalic cranium, cranial base, and facial structures would undoubtedly enrich our knowledge of the normal processes of growth and development of these regions.

Summary

Posteroanterior and lateral cephalograms of twenty microcephalics were used to compute cranial volume and study cranial base alterations. Statistical analyses of the data indicated various significant relationships which led to the following conclusions: 1) Cranial volume estimation *in vivo* using cephalograms is a practical method for classification of microcephalics. 2) A highly significant shortening of the cranial base exists in microcephalics. The posterior cranial base (S-Ba) was most significantly reduced in size, the external anterior cranial base (N-S) next and then the internal anterior cranial base (S-F). 3) A high correlation exists between the cranial circumference and the cranial volume. A moderately positive correlation exists between the length of the cranial base (N-Ba) and the cranial circumference. No correlation was found between the cranial volume and the cranial base. 4) A highly significant correlation exists between the thickness of the frontal bone (N-F) and the distance between the cribriform plate and the N-S line. A similarly positive correlation was found between the eth-N-S line and internal cranial base angulation (eth-S-Ba). 5) The distance from the orbital roof of the N-S line is very close to the normal range. The thickness of the frontal bone in subjects under the age of twenty is normal. Over twenty years of age, the frontal bone appears somewhat below average in size.

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